

Carotid endarterectomy in an asymptomatic patient with contralateral agenesis of the internal carotid artery

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A 64-year-old man was referred to the vascular surgery department with an incidental finding of right internal carotid artery stenosis during thyroid ultrasound imaging. Carotid artery color duplex revealed an ulcerated type III plaque producing stenosis of the right internal carotid artery of 70% and agenesis of the left internal carotid artery. Further imaging with computed tomography angiography confirmed the initial findings and excluded cerebral vascular malformations and ischemic cerebral infarcts. The patient was treated with endarterectomy of the right internal carotid artery with elective shunting and synthetic patch closure. (*J Vasc Surg Cases* 2015;1:254-7.)

Congenital abnormalities of the internal carotid arteries (ICAs) in the form of agenesis, aplasia, or hypoplasia, refer to a group of dysgenesias during embryonic development. Prevalence in the general population is rare, <0.01%.¹⁻³

Patients are mostly asymptomatic, and the abnormality is often an incidental finding during imaging performed as the result of other cerebrovascular events. Rarely, they can be discovered during cervical ultrasound imaging.^{1,2,4-6} We report the case of a 64-year-old man with agenesis of the left ICA who was treated with carotid endarterectomy (CEA) of the right ICA for an asymptomatic stenosis of 70% with a type III ulcerated plaque. The patient consented to publication of this report.

CASE REPORT

An asymptomatic 64-year-old man was referred to the vascular surgery department on the basis of right ICA stenosis of 70% found during thyroid gland ultrasound screening. Stenosis of the left ICA was characterized as preocclusive, and the left common carotid artery (CCA) was characterized as hypoplastic. The left vertebral artery was oversized, and flow was significantly higher than in the left CCA.

The patient's medical history revealed familial hypercholesterolemia and hypothyroidism. He was taking oral levothyroxine therapy (85 µg once daily) and acetylsalicylic acid (100 mg once daily) but could not tolerate statin therapy. Initial laboratory

results were within normal reference ranges, accompanied by a normal chest X-ray and a normal electrocardiogram. A previous cerebral computed tomography scan was normal.

Color duplex imaging of the carotid arteries was repeated by an affiliated specialist, and the initial finding of right ICA stenosis of 70% was confirmed, with an ICA peak systolic velocity of 230 cm/s and a ICA-to-CCA peak systolic velocity ratio of 4. The atherosclerotic plaque was characterized as type III (grey scale median = 40), and a significant ulcerated area was noticed (*Fig 1*). Left ICA agenesis was diagnosed. The left CCA was hypoplastic, and the left vertebral artery was significantly dilated. Computed tomography angiography (CTA) confirmed all latter color duplex findings and demonstrated complete absence of the cervical, petrous, and cavernous left ICA (*Fig 2*).

CEA of the right ICA under general anesthesia was performed with selective use of a Pruitt-Inahara Carotid shunt (LeMaitre Vascular Inc, Burlington, Mass) and synthetic Dacron patch (Intervascular, Le Ciotat, France) closure, with no neurologic deficit (*Fig 3*). In our department, we selectively use a shunt for every CEA with contralateral ICA occlusion, and the ICA aplasia in this patient was considered an occlusion. The patient was discharged on the second postoperative day and remains asymptomatic.

DISCUSSION

The first report of ICA aplasia in a cadaver was by Tode⁷ in 1787. It was very later, in 1954, that the first angiographic images of ICA agenesis were acquired by Verbiest,⁸ and Handa et al⁹ reported the associated absence of the bony carotid canal in 1980.

Absence of ICA is considered a rare congenital anomaly found in <0.01% of general population, and bilateral absence of ICA is an even smaller percentage of this group of patients. Dysgenesias of the ICAs are classified into agenesis, aplasia, and hypoplasia. We refer to agenesis as total failure of the ICA formation, aplasia when a precursor is present in the embryo and is represented by a remnant such as a fibrous band, and hypoplasia when a small-caliber ICA is present.³

The etiology of ICA formation anomalies is not known. The ICA forms from the third aortic arch and the CCA from

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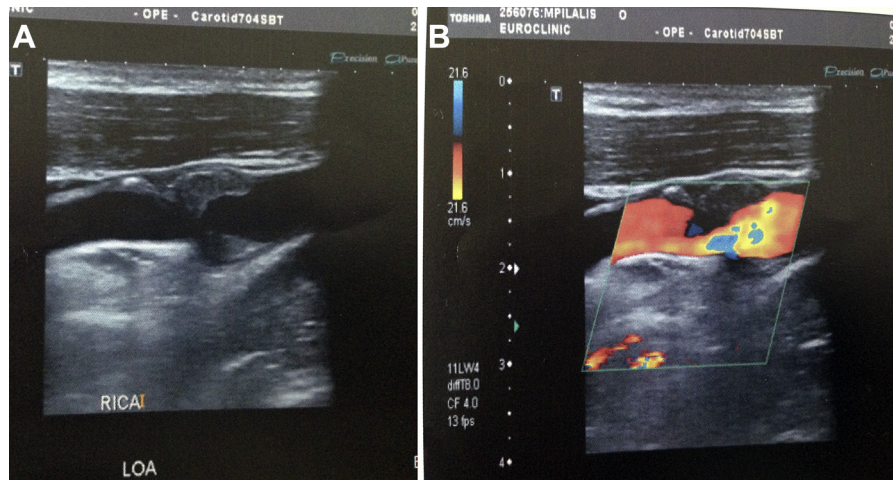


Fig 1. Color duplex findings. **A**, An ulcerated type III atherotic plaque in the orifice of the right internal carotid artery (RICA). **B**, Turbulent flow due to a 70% stenosis in the RICA orifice.

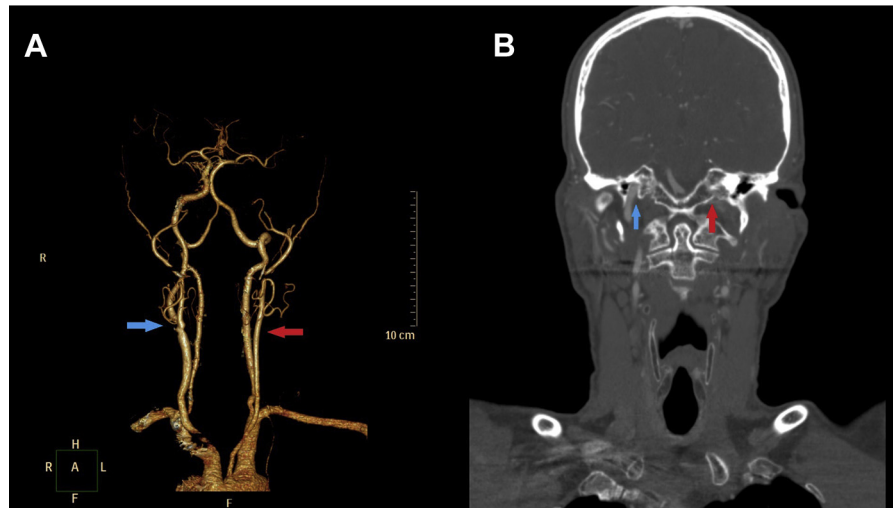


Fig 2. **A**, Three-dimensional reconstruction of a computed tomography angiography (CTA) image. The left common carotid artery (CCA) is hypoplastic (red arrow), the left vertebral artery is dilated, and the right internal carotid artery (ICA) stenosis can be noted (blue arrow). **B**, The right ICA can be seen in the carotid duct (blue arrow). On the left side, there is absence of the carotid duct (red arrow).

the ventral aortic root between the third and fourth aortic arches. Lasjaunas et al¹⁰ were the first to propose the division of the ICA into six segments: cervical, petrous, vertical cavernous, horizontal cavernous, clinoid, and cisternal. Their development follows the development of the corresponding embryonic arteries: ventral pharyngeal, hyoid, mandibular, primitive maxillary, trigeminal, dorsal ophthalmic, and ventral ophthalmic, respectively. Every part is independent, with a specific course and limit, and because each one forms independently, anomalies may result in segmental-only agenesis. ICA formation is completed at the fourth embryonic week, and the carotid canals form between the fifth and sixth embryonic weeks.^{3,11} The presence of an ICA is

mandatory for the formation of a carotid canal, and so ICA agenesis results in concomitant carotid canal agenesis. The size of carotid canals is reduced proportionally in aplasia and hypoplasia.¹² There are several possible collateral pathways to counteract for the decreased blood supply in the case of ICA agenesis, including the posterior cerebral circulation through the posterior communicating arteries, enlarged branches of the internal maxillary artery, intercavernous collaterals, or even persistent fetal circulation.^{1,12}

Dilatation and accompanying increased blood flow through the vertebrobasilar system, the anterior cerebral circulation, or through the external carotid arteries, cover the requirements for cerebral perfusion. It has been suggested

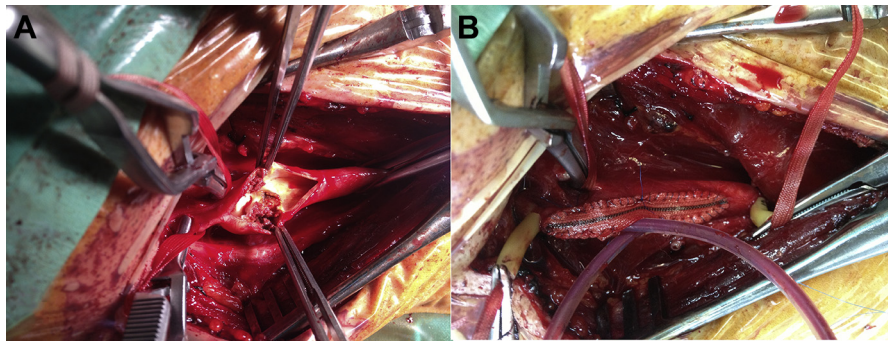


Fig 3. Intraoperative images. **A**, Ulcerated atherotic plaque. **B**, Synthetic Dacron patch (Intervascular, Le Ciotat, France) closure with the use of selective shunting (LeMaitre Vascular Inc, Burlington, Mass).

that in cases of unilateral ICA agenesis, the dilated contralateral ICA substitutes through the anterior communicating arteries the blood flow in the agenetic segment¹³ and that in bilateral agenesis, the increased blood flow through the vertebrobasilar system replaces the blood flow in the agenetic segments.³

In our patient, the ipsilateral vertebrobasilar system was dilated, providing increased collateral blood flow, as depicted in the findings of the color duplex imaging and the CTA. Hemodynamic stress placed on the vessels due to increased flow results in the development of intracerebral aneurysms.¹ Their prevalence is 2% to 4% in the general population and 24% to 34% in patients with ICA agenesis.¹³ Hypertension is considered an aggravating factor.^{2,4}

Aneurysms are most frequently located in the anterior communicating artery¹ but can also be found at the first part of the subclavian artery,¹⁴ posterior communicating, posterior cerebral, or the basilar artery.¹⁵ Other findings are cerebral arteriovenous malformations, hypopituitarism, neurofibromas, interruption of the aortic arch, aberrant origin of the ophthalmic artery, CCA agenesis or hypoplasia, basilar artery aplasia with nasopharyngeal angiofibroma, corpus callosum agenesis, and congenital temporomandibular joint ankyloses.¹⁶ ICA agenesis is also met in patients with PHACE (Posterior fossa, Hemangioma, Arterial lesions, Cardiac abnormalities/aortic coarctation, Eye abnormalities) anomaly.¹⁴ Tsuruta and Miyazaki¹⁷ classified three collateral circulatory configurations, and Lie¹⁸ summarized six basic patterns. Therefore, intracranial artery imaging in cases of ICA dysgenesis is mandatory.

Patients with ICA agenesis rarely report symptoms. Cerebral perfusion is adequate due to collaterals; therefore, only one report of a symptomatic child has been published.¹ Transient ischemic attacks, stroke, and thromboembolic episodes present in advanced age as atheromatous disease progresses later in the collateral vessels. Horner syndrome, cranial nerve palsies, headaches, migraines, and seizures have also been reported.¹¹ Subarachnoid or intracerebral hemorrhages are uncommon complications from associated cerebral aneurysms or arteriovenous malformations.¹⁴

Color Doppler ultrasound imaging has high sensitivity for the detection of ICA agenesis but is operator dependent

and cannot identify the abnormality as a whole. Distal parts of the ICA are not always accessible, and the skull base cannot be assessed. CTA and magnetic resonance angiography have the advantage of depicting the CCA, ICA, and external carotid artery throughout their course from the aortic arch to the base of the skull and can also identify the carotid canal. Modern visualization software allows for three-dimensional reconstructions and subtractive imaging. CTA is faster and widely available but has the disadvantage of ionizing radiation. Digital subtraction angiography produces false-negative results because the acquired image is similar to an ICA occlusion.^{19,20}

Conditions with severely narrowed or occluded ICA are atherosclerosis, arteritis, arterial dissection, fibromuscular dysplasia, moyamoya disease, and sickle cell disease.¹⁴ The presence of vessel stump, thickened arterial wall, and the atheromatous plaque or thrombus can direct to a proper differential diagnosis between agenesis and occlusion. Absence of the carotid canals helps further discrimination between acquired stenosis, hypoplasia, and agenesis. In our patient, the left carotid canal was absent.

In cases of CEA of the contralateral ICA, the decision whether to shunt the patient can also be made using electroencephalogram or stump back-pressure criteria, because no guidelines currently exist due to the rarity of the condition. Indications for contralateral CEA are the same as in the general population.

CONCLUSIONS

Agenesis, aplasia, and hypoplasia of the ICA are rare vascular congenital defects, mostly asymptomatic and undetected, but their recognition is of major clinical interest. In addition to associated cerebral aneurysms, these dysgenesias have important implications during CEA, neurosurgery, and in the setting of cerebral embolic events.

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